Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis in a Patient With Antiphospholipid Syndrome

We report a case of idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) in a patient with antiphospholipid syndrome.

Report of a Case. A 76-year-old white man had slightly blurred vision and visual acuity of 20/30 OU. Slitlamp examination showed anterior chamber inflammation and rare cells in the anterior vitreous. Funduscopic examination (Figure 1) showed hard exudates and intraretinal hemorrhages. Fluorescein angiography (Figure 2) emphasized the multiple arteriolar aneurysms, most prominent at bifurcations. Leakage from and staining of both arterial and venous vessels as well as the optic nerve is best appreciated in the right eye.

His history was significant for a seizure disorder treated with phenytoin sodium for 25 years and coronary artery disease treated with a stent 1 year prior to our examination. He has lived in Kenya, Uganda, and the West Bank and has been treated for malaria. He also had occipital-lobe stroke 16 years earlier, which was thought to be related to a patent foramen ovale. A patent foramen ovale was not noted on a transthoracic echocardiogram. He has been receiving warfarin sodium continuously since the stroke, with a goal international normalized ratio of 2.5 to 3.5.

A review of his previous ophthalmology records showed at least a 7-year history of vitritis, retinal vasculitis, and hard exudates more prominent in his right eye. During this time, no ocular treatment was performed.

Laboratory and imaging tests with results within normal limits included complete blood cell count, metabolic panel, liver enzymes, angiotensin-converting enzyme, erythrocyte sedimentation rate, hepatitis B and C antibodies, fluorescent treponemal antibody absorption, rapid plasma reagin, Bartonella henselae and Bartonella quintana IgG and IgM, Toxoplasma gondii IgG, and...
tinuclear antibody, human immunodeficiency virus, Lyme disease, human T-lymphotropic virus types I and II, antiproteinase 3, antitymelyperoxidase, chest computed tomography, urinalysis, serum protein electrophoresis, and carotid Doppler ultrasonography. T. gondii IgG was positive and rheumatoid factor was slightly elevated at 23 IU/mL.

Antiphospholipid syndrome laboratory test results included the following (the 2 values were obtained 12 weeks apart): activated partial thromboplastin time, 40.2 and 49.2 seconds (reference range, 20.0–37.6 seconds); dilute Russell viper venom time, 68.2 and 71.8 seconds (reference range, <49.0 seconds); hexagonal phospholipid neutralization, 16.7 and 5.4 seconds (reference range, <8 seconds); anticardiolipin IgG, 77 and 118 GPL/mL (high positive, >50 GPL/mL); anticardiolipin IgM, 37 and 29 MPL/mL (high positive, >30 MPL/mL); anti–β2–glycoprotein I IgG, 117 and 53 SGU/mL (positive, >20 SGU/mL); anti–β2–glycoprotein I IgM, 68 and 40 SMU/mL (positive, >20 SMU/mL); antiphosphatidylserine IgG, 4 and 88 GPS/mL (high positive, >50 GPS/mL); and antiphosphatidylserine IgM, 21 and 52 MPS/mL (high positive, >50 MPS/mL).

Comment. Our patient demonstrates the 3 major criteria of IRVAN: retinal vasculitis, aneurysmal dilations at arterial bifurcations, and neuroretinitis. He falls into the stage 1 category. However, our patient differs from a typical patient with IRVAN in that he is male, is older than 50 years, and has a probable associated systemic disease.

Our patient, with a history of stroke and medium to high titers of 2 types of antiphospholipid antibodies (anticardiolipin and anti–β2–glycoprotein I) on 2 separate occasions 12 weeks apart, would definitely be diagnosed as having antiphospholipid syndrome. Our patient is also likely positive for lupus anticoagulant as well, but the interpretation is more controversial in the setting of oral anticoagulation. Results of lupus anticoagulant screening tests (activated partial thromboplastin time and dilute Russell viper venom time) were positive on both occasions, but the result on the confirmatory test, hexagonal phospholipid neutralization, was positive on 1 occasion.

To our knowledge, this is the first reported case of a patient with both antiphospholipid syndrome and IRVAN. The presence of both antiphospholipid syndrome and IRVAN in our patient could have occurred by chance alone. However, an association is plausible as both conditions affect the vasculature and both are uncommon. When evaluating a patient with IRVAN, one should consider the possibility that lupus anticoagulant and antiphospholipid antibodies are present, especially if the patient has a history of nonocular vascular thrombosis.

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